

## *Case Report*

# Rapid idiopathic osteolysis of the shoulder

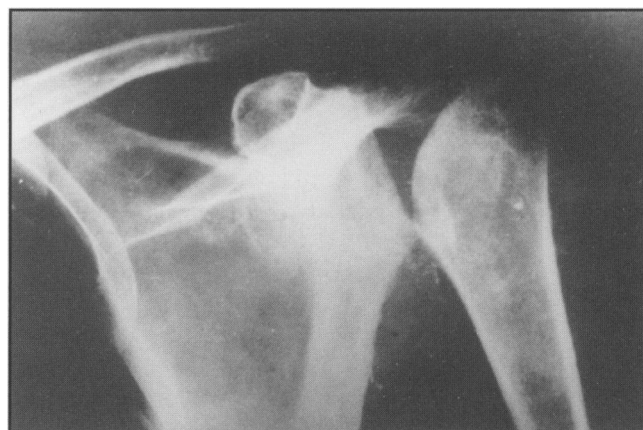
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This case reports a patient with idiopathic osteolysis of the shoulder. The clinical, radiographic, and histological features are presented. Complete destruction of bones without known causes was observed only sixteen months after onset of symptoms. The authors believe this to be the first case of such rapid progression of this disease.

Idiopathic osteolysis (Gorham's disease) is an extremely rare occurrence. The etiology of osteolysis is as yet unknown. A history of minor trauma may be a factor. Bones begin to resorb, partially or completely as a localized erosion, and from this, slow centrifugal absorption of a large piece of the skeleton takes place. The osteolytic process may halt spontaneously after years of progression. Even should it, remineralisation typically is not seen and pathological fractures that occur in patients who have idiopathic osteolysis do not heal.<sup>1-13</sup> Bones are usually replaced with fibrous tissue rich in proliferating capillaries and dilated vessels with no symptoms or signs of acute inflammation.<sup>1</sup> In late stages of the condition, vascular proliferation appears to regress, which leaves a largely fibrotic stroma in its place.<sup>4</sup> The results of osteolysis are deformity and impaired function. Some patients die as a direct result of massive osteolysis.<sup>9</sup> The purpose of this paper is to point out the course of severe rapid progression of idiopathic osteolysis.

**CASE REPORT** We report the case of a 54-year-old man who complained of pain and stiffness in the left shoulder of gradual onset over six months. The pain was not severe, it varied considerably but was becoming worse, leading to restricted movement of the shoulder. We could not find history of any trauma or bone disorders neither with him nor with anybody from his close family. On the first examination, active movements of the shoulder were grossly reduced,



*Figure 1*

but passive movements were exaggerated and painful. Movements in the elbow and the wrist of ipsilateral side were good as well as grip strength. There was absence of any palpable soft tissue mass. X-rays taken at the time showed atypical formation of humeral head, with areas of absorption in lateral part of clavicle involving acromioclavicular joint and lateral part of acromion (Fig 1). The results of laboratory investigation (sedimentation, full blood count, serum calcium, liver test, RA factor, Nelson's test, antinuclear antibodies) were in the normal range as well as renal function and angiography. On biopsy we found fibrous tissue rich in proliferating capillaries with huge perivascular mononuclear cells. He received a course of physical therapy with some pain decrease. The

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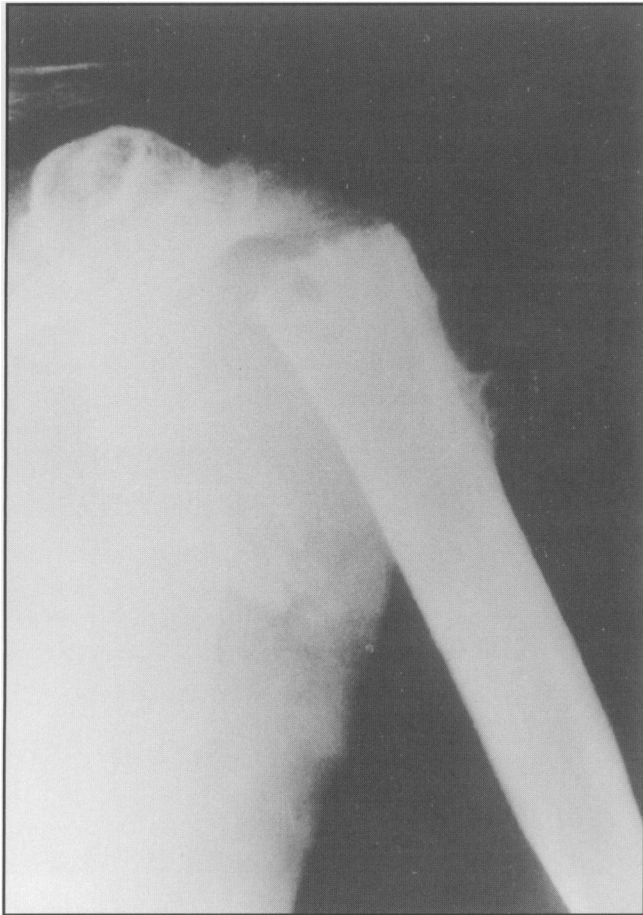


Figure 2

patient returned after 4 months because of exacerbation of pain, which became worse over the outer side of the left arm, top of the shoulder and the scapular region. He complained on losing range of movement and strength. Control radiographs showed absence of the humeral head on left side with destruction of lateral margin, glenoid cavity and apex of the scapula. We observed progression of osteolytic changes on the clavicle too. There was no evidence of new bone formation (Fig 2).

In the next month we registered rapid progression of osteolysis. Sixteen months after onset of the symptoms, X-rays showed that the shoulder completely disappeared with dispersed pieces of necrotic bones in adjacent soft tissue (Fig 3). In this phase, progression of osteolysis spontaneously stopped. The patient is still under observation. Regarding the treatment, he was given orthotic appliances as well as exercises to strengthen the shoulder and arm muscles. In spite of this there are activities that he can not perform because of the instability and lack of strength in the involved shoulder.



Figure 3

## DISCUSSION

The etiology of idiopathic osteolysis is still unsolved and the prognosis remains unpredictable. The localization of osteolysis may vary as may its spread to adjacent bones. This condition causes considerable concern in the early months, and essentially, it is a diagnosis per *exclusionem*. First one must exclude malignant osteoclastic tumours and inflammatory disorders of bone. Secondly, arterial vascular diseases, neurogenic arthropathies, and other neurological diseases must be ruled out. Thirdly, the post-traumatic osteolyses must be considered because this can present a similar picture to idiopathic osteolysis. Torg *et al*<sup>13</sup> classified osteolysis into four types: idiopathic multicentric osteolysis with dominant transmission, idiopathic multicentric osteolysis with recessive transmission, idiopathic non-hereditary multicentric osteolysis with nephropathy, and Gorham's massive osteolysis. Macpherson *et al*<sup>8</sup> added a fifth type, namely the Winchester syndrome. Because of the rarity of this disease, the assessment of any method of treatment is difficult and unpredictable. Until

now, many methods have been proposed with varying success,<sup>4, 5, 10, 11</sup> but effective therapy is still being sought. Since 1838, when Jackson reported the first case of idiopathic osteolysis,<sup>5</sup> only a few other cases have been reported in the literature under a wide variety of names which include acute spontaneous absorption of bone, massive osteolysis, phantom bone,<sup>4</sup> and disappearing or vanishing bones.<sup>9</sup> This case seems to be the eighth with involvement of the scapula. The progression of the condition varies, but a duration of less than several years is rare.<sup>1-9</sup> The fastest progression of idiopathic osteolysis was described by Kareem *et al* in 1994.<sup>6</sup> They described massive osteolysis of pelvis the pelvic girdle over a three-year period. In the case we present here, complete destruction of the shoulder-bones was observed only sixteen months after the onset of symptoms. To the authors' knowledge, such rapid progression of massive osteolysis has not been previously reported. Further new cases may well provide insight into the clinical and pathophysiological features of this disease.

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